

Dupuytren's Contracture—Etiology

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■ *Although Dupuytren's contracture was first described more than 300 years ago, the cause is not yet certainly known. It is only agreed that the disease process affecting the palmar fascia is most commonly found in older-aged males of Caucasian descent.*

Conservative or non-surgical treatment is usually ineffective. Surgical treatment includes fasciotomy, partial fasciectomy and total fasciectomy. Fasciotomy is indicated for localized disease or for patients with medical conditions which preclude more extensive operation. Subtotal fasciectomy with or without skin graft is the most commonly employed procedure. Total palmar fasciectomy is seldom indicated. Improved function of the hand can be anticipated after adequate surgical intervention.

and Principles of Treatment

PULVERTAFT AND REID in 1963 stated: "It is humbling to reflect that although Dupuytren's disease has been known since the description by Plater (1614), Astley Cooper (1822) and Guillaume Dupuytren (1832) the cause remains obscure and there is no general agreement as to the ideal method of surgical treatment.^{1,3} Dupuytren, although historically preceded by Clive in 1808 and Cooper in 1822 in the recognition of the fascial origin of the disease, was the first to perform an operation for relief of the contracture.^{4,5}

Etiology

The exact cause of Dupuytren's contracture is unknown. Predisposing factors which are universally accepted are age, sex, race and heredity.⁴ Dupuytren's contracture is observed most often in the fifth through seventh decades. There are occasional occurrences in the third and fourth decades and very rare occurrences in the second.

Dupuytren's contracture is a disease of men. Most reported series indicate that the disease occurs seven to eight times more frequently in men than in women.

Most investigators agree that this disease is much more common in the non-pigmented races and is uncommon in Chinese.

Larsen noted that there is a definite hereditary tendency toward the development of the disease and states that European writers frequently report as many as one-third of their patients with Dupuytren's contracture have a positive family history.⁵

The role of trauma in the development of this disease is one of the major points of disagreement among surgeons treating this problem. Clarkson in 1961, in a survey of numerous authorities, found that most opposed the view that a direct relationship existed between trauma and Dupuytren's disease.⁶ There was some opinion, however, that trauma to the medial side of the palm followed by fixation of the digits in flexion might represent a causal relationship to the precipitation or aggravation of the disease. Many historical studies, including those of Early, Herzog and Hueston, failed to show any occupational connection with chronic trauma.⁷⁻⁹ In contrast, there has been a significantly high, but as yet unexplained, incidence of Dupuytren's disease noted among brewery workers, alcoholics, and epileptics.^{9,10}

The Palmar Fascia^{11,12}

The palmar aponeurosis forms the volar wall of a space that contains the flexor tendons, the lumbrical muscles and the vessels and nerves of the palm. Proximally it is joined to the palmaris longus tendon, or if the tendon is absent, to the forearm fascia. In the distal half of the palm septa pass into the depths of the palm between the tendons of the adjacent fingers. These vertical septa join the deep palmar fascia and thereby form arcades. Eight such arcades are formed with each alternate tunnel transmitting a lumbrical muscle and digital vessels and nerves; each of the other four tunnels transmit a pair of flexor tendons.

Near the heads of the metacarpals a transverse ligament is prominent which spans the space between the four main longitudinal bands. These

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Figure 1.—Hand of 45-year-old man with decided flexion deformity of the little finger. Note the prominent knuckle pads over the proximal interphalangeal joints of the ring and middle fingers. The fixed flexion contracture of the proximal interphalangeal joint of the little finger was arthrodesed in 45° of flexion after excision of the diseased fascia in the palm and digit.

longitudinal palmar bands pass into the superficial digital fascia and the vertical septa pass into the deep digital fascia. These fibers from the palm which pass into the fingers ultimately attach to the sides of the proximal and middle phalanges and thereby account for the proximal interphalangeal joint contracture which may occur.

Clinical Features

The Dupuytren's lesion may become manifest in a variable manner; it may be noted to be only a small palmar nodule or it may be complete contracture of the ring and little fingers which are firmly opposed to the palm. The palmar nodule represents the first manifestation of the disease in the palm and, although progression is usually noted beyond this stage, it is entirely possible that the nodule may not progress to involve the remainder of the palm or digital fascia. If progression occurs, thickening of the palmar fascia on the medial side of the hand is noted with associated flexion contracture at the metacarpal phalangeal joint. Further progression of the disease is often associated with a flexion deformity at the proximal interphalangeal joint. The ring finger is most commonly involved with the little finger being the next most commonly involved.^{13,14}

Dupuytren's knuckle pads may be noted over the dorsal aspect of the proximal interphalangeal joints. Skoog noted their presence in 44 percent of patients with Dupuytren's contracture.¹⁵ These nodular subcutaneous masses are illustrated in Figure 1.

Progression of the disease may be variable and



Figure 2.—(A) The patient, a 70-year-old man, had pronounced flexion deformity of the little and ring fingers. (B) Simple fasciotomy under direct vision produced remarkable improvement. The longitudinal incision provided for early motion without separation of the wound.

the surgeon must recognize this fact. Moreover the patient should be told, before any surgical intervention, that the disease which has affected his hand or hands is one that is characterized by chronicity and recurrence. This is easily understood in that the palmar fascia is an extensive tissue involving the entire palmar area with extensions into the proximal phalanx of each of the fingers. Although partial excision of the fascia may be performed, residuals of palmar fascia remain and may represent sites for future involvement with disease. Hueston noted that in spite of complete excision of palmar fascia and abnormal tissue, recurrence is common in patients who were operated upon during the phase of rapid progression of the disease.⁹ The fact that nodules or recurrent Dupuytren's disease may develop in an area previously operated upon indicates, in Hueston's opinion, that these recurrent nodules could not have arisen from any preexisting anatomical fascial bands but developed from the normal fibro-fatty tissue remaining in the region.

Treatment

Larsen observed that non-operative treatment of Dupuytren's contracture with local injections of pepsin, fat solutions, fibrinolysin, copper sulfate, and pancreatic extract and cortisone have had no effect upon the disease. Treatment with oral tocopherol may produce temporary softening of the contracture but the progressive course of the disease is not altered.¹⁶ Although Finney suggested a combination of preoperative irradiation and surgery for advanced cases, Larsen observed that the

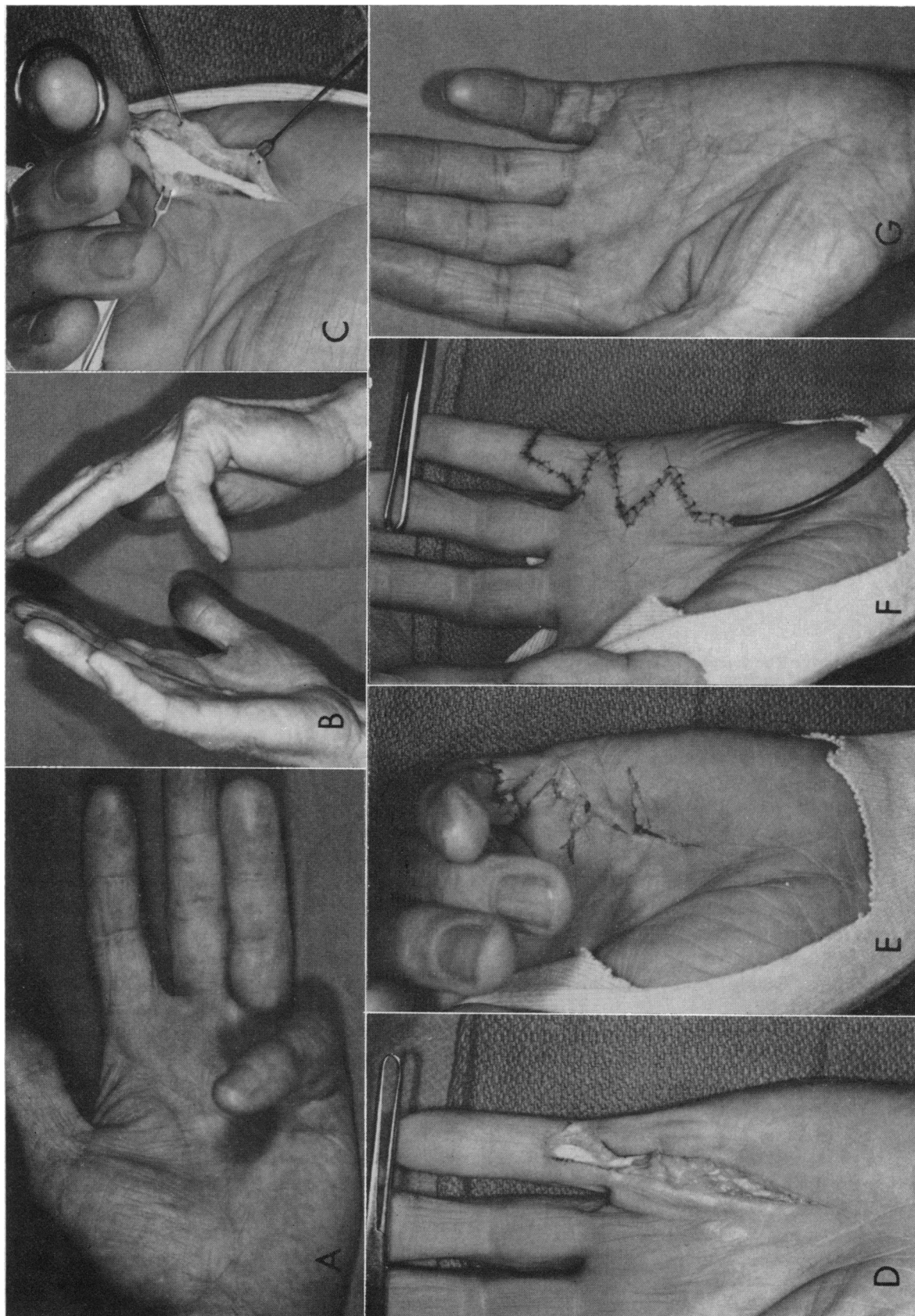


Figure 3.—(A and B) Hand of 50-year-old man with isolated disease involving the fifth ray. A single "cord" of hypertrophied fascia passed from the proximal palm into the proximal interphalangeal joint of the little finger. Maximum extension is demonstrated in B. (C) A longitudinal incision was used to expose and excise the diseased fascia. (D) Note the normal fat and underlying flexor tendons after excision of the fascia. (E and F) Z-plasty closure was performed as noted. The drain (F) is considered essential. (G) The cosmetic and functional result is considered to be good.

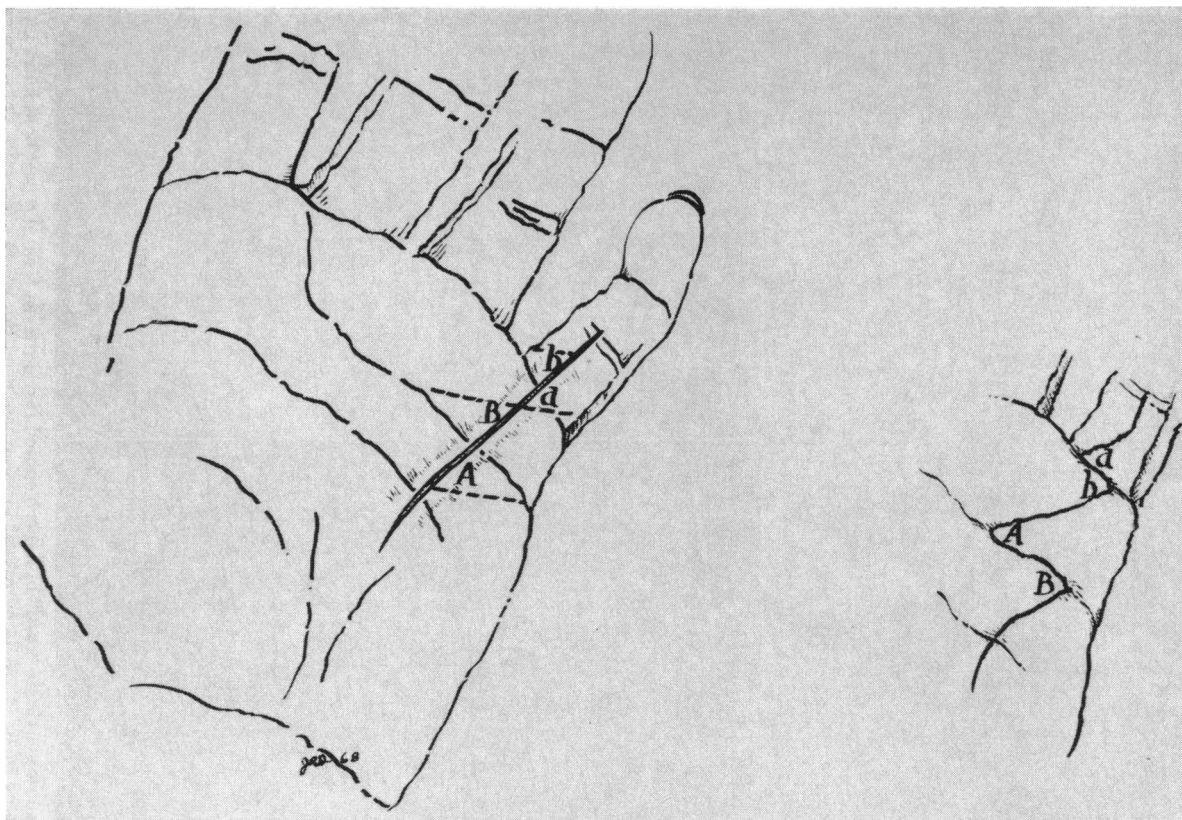


Figure 4.—Z-plasty: The longitudinal approach provides for direct exposure with a minimum of soft tissue undermining. A longitudinal incision in this area, if allowed to heal, would produce recurrent flexion deformity because of the longitudinal scar which crosses two flexion creases at right angles. Accordingly, the Z-plasty is used to break up this line of tension. In addition, the Z-plasty technique provides for longitudinal lengthening of contracted skin.

results were less satisfactory than might be expected from surgery alone.^{16,17}

The majority of physicians dealing with this process agree that the most effective treatment is surgical. Operation is by no means indicated in all cases of Dupuytren's disease. When the contracture interferes with function or occupational requirements, surgical treatment is indicated. The aims of operation are to correct or partially correct the relatively fixed flexion contractures involving the affected digits.

Unfortunately it is impossible to forecast accurately the probability of progression of the disease. Mild disease which remains static does not warrant operation. However, such early operation is technically easier and the results are better. Conversely, long-standing contracture is often associated with permanent joint changes and a perfect result is unlikely. A few months' observation may answer the question regarding the need for operation. If the contracture progresses rapidly, surgical correction is recommended. If it remains static (and the deformity is mild) operation is not indicated.

The operation must be designed to fit each patient. Age, general health and functional needs must be considered. Postoperative joint stiffness is the major complication of operative treatment,¹⁸ particularly if arthritic changes are already present. Similarly, a thick broad hand does not withstand extensive surgical intervention and is prone to postoperative stiffness.

The three main forms of surgical treatment are fasciotomy, partial fasciectomy and a complete or radical fasciectomy. Fasciotomy may be the procedure of choice in older persons whose general medical condition precludes more extensive operation. It can be performed under local anesthesia and is a practical answer for limited and relatively isolated bands of contracted fascia. Fasciotomy may also be used as a preliminary procedure in severe contractures in preparation for more radical procedures. The surgeon may be surprised to note the pronounced functional gains from this relatively simple procedure. Longitudinal incisions are advocated, since they permit early motion without gaping of the wound. The contracted band is iso-

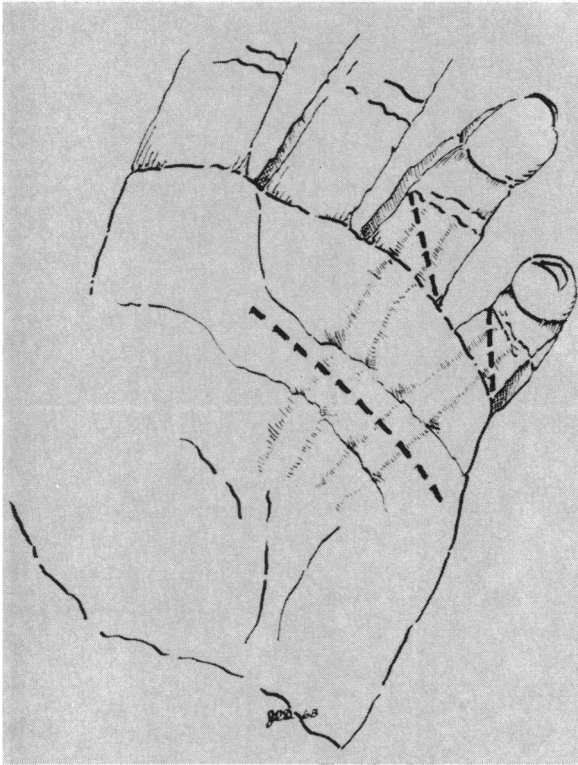


Figure 5.—The transverse palmar incision is used if extensive disease is noted in the palm. It is supplemented with digital incisions as needed.

lated and sectioned under direct vision with a scalpel. Blind sectioning of such bands may result in inadvertent transection of nearby vessels or nerves (Figure 2).

Partial fasciectomy is the most commonly employed technique and involves excision of the diseased fascia in the palm and fingers. It must be observed that the contracted fascial bands originate in the palm but extend into the proximal phalanx and across the flexor and lateral aspect of the proximal interphalangeal joint. Surgical exposure must therefore be directed toward the palm and the proximal phalanx. If a single longitudinal band is present, it may be exposed through a longitudinal incision which is closed by a Z-plasty (Figures 3 and 4). If the disease process is more extensive, a transverse palmar incision and oblique digital incision may be used for good exposure (Figure 5). A surgical tenet as regards incisions is to design the incision so that all diseased fascia can be removed with a minimum of dissection. Obviously, each hand must have incisions which meet its particular requirements.

It is the author's opinion that, anatomically, total fasciectomy is seldom indicated and that the

surgical approach to Dupuytren's disease should be that of relatively total excision of all diseased palmar fascia including digital extensions. This may result in relatively localized operation with the understanding that recurrences may occur in areas which were not previously involved. It must be remembered that the remaining fascia has potential for hypertrophy.

Cases of long-standing contracture are often associated with contracture of the skin in the palm. In addition, some cases of severe Dupuytren's disease manifest infiltration of the skin so that the disease process cannot be excised without leaving a very thin layer of skin in some areas. If extremely thin or excessively traumatized skin remains following fasciectomy, it may be necessary to replace the skin by a skin graft. On many occasions, after complete excision of the diseased fascia a large gaping wound will be observed in the palm or digits with passive extension of the fingers. Primary closure of these wounds would result in a considerable flexion deformity of the fingers and it is the author's opinion that this flexion deformity can be prevented by the insertion of a full thickness skin graft into the defects (Figure 6).

During dissection it is extremely important to carefully identify the digital arteries and nerves in order to avoid injury to them. This is especially important in the proximal phalanx where the nerves may be very close to the digital fascial bands. Extensive undermining should be avoided. The studies of Conway have demonstrated that the arterial supply of the palmar skin is in a mosaic fashion and that skin necrosis is likely to follow the wide and indiscriminate undermining in the palm.¹⁹ It is the author's custom to drain all the operative wounds by the use of a sterile No. 8 (French) red rubber catheter. The tip of the catheter is passed through the proximal portion of the incision and down to the distal area of dissection. The catheter is relatively small and is well tolerated by the tissues. After closure of the skin, the catheter is coiled on the flexor surface of the wrist and can easily be removed in 48 hours without disturbing the primary compression dressing.

Certain digits, especially the little finger, often are associated with extreme degree of contracture and the need for amputation of this digit is often entertained. This is especially true when the deformity is long-standing and is fixed. Although an amputation is easily performed and the cosmetic

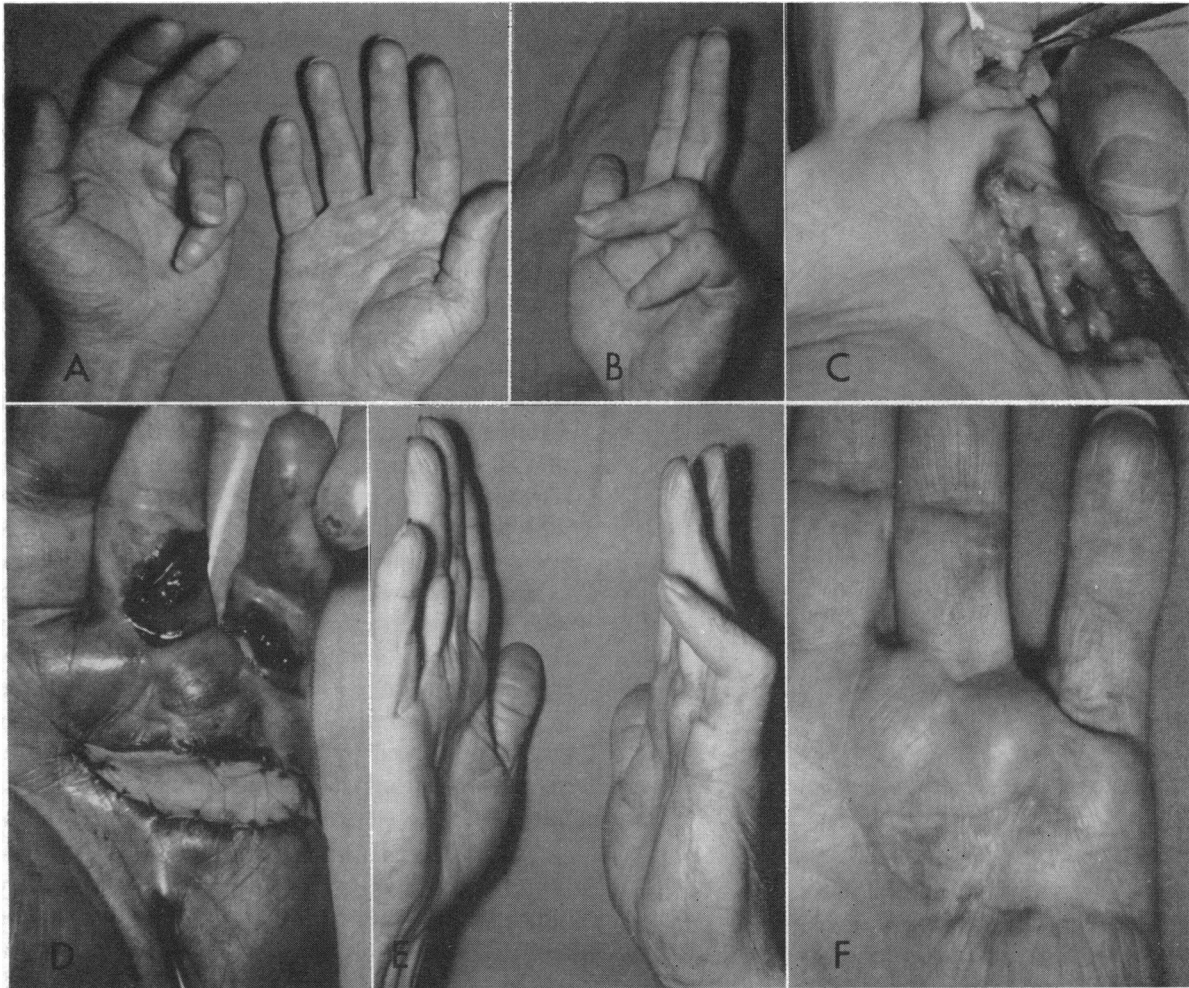


Figure 6.—The patient was a 45-year-old man. (A and B) Note the decided flexion deformity of the ring and little fingers of the left hand. (C and D) Palmar and digital incisions were used to excise all diseased fascia. The resultant skin defect in the palm required a full thickness skin graft. A similar graft was placed in the defect in the little finger. Primary closure of these wounds would have resulted in early recurrence of the flexion deformity. (E and F) The result is considered to be very satisfactory.

and functional results are not unreasonable, many surgeons hesitate to perform this practical solution. Amputation is especially recommended in long-standing cases of fixed flexion contracture of 90 degrees or more which involve both the proximal interphalangeal and the metacarpal phalangeal joints.

Fixed flexion contracture of 90 degrees or more of the proximal interphalangeal joint is a most difficult problem to resolve. Long-standing contracture is associated with contracture of all surrounding soft tissues and even extensive dissection and surgical release cannot effect permanent changes. Forced extension at the operating table, if associated with subluxation of this joint, predicts recurrence of the deformity in the author's experience. When such irreparable joint changes are encoun-

tered, the best and most lasting result is arthrodesis of the joint in 45 degrees of flexion. Two cases of this type were encountered in this series. The methods used by the author in fifty cases were as follows: Fasciotomy, 3; partial fasciectomy, 40; partial fasciectomy with skin graft, 6; amputation of digit, 1.

Local or distant pedicle flaps may be used to provide coverage when a large defect results from adequate excision of the diseased palmar fascia and skin.²⁰ The filleted skin of an amputated little finger certainly provides an ideal flap for this purpose. Less ideal are the chest or abdominal pedicle flaps and the least desirable is the local flap which is obtained by rotating a dorsal hand flap into the palm.

The dressing applied immediately following op-

eration is of great importance. One of the most disastrous complications which can occur following excision of Dupuytren's contracture is the occurrence of a hematoma in the palm. Such a hematoma results in devitalization and compromise of the blood supply to the palmar skin and may result in necrosis of the skin. In the author's experience, a hematoma can be prevented by pre-closure hemostasis and adequate drainage with the previously described catheter. In addition, the dressing must be of a moderate compression type with several fluffs of gauze placed on the palm and on the dorsal surface of the hand and fingers. The dressing extends from the fingertips to the wrist. The thumb is allowed to remain out of the dressing and acts as a stabilizing post to prevent migration of the dressing. Sponge rubber or a synthetic variant of at least one-inch thickness is placed in a "pancake" fashion on the palmar and dorsal surfaces of the hand and fingers. This is firmly wrapped into place with bias-cut stockinette bandage which further adds to the gentle compression. The author is of the opinion that the fingertips should be splinted in this fashion in as much extension as the soft tissues will tolerate.

The objectives of treatment include correction of a flexion contracture and temporary extension of the fingers for seven to ten days has been observed not to result in permanent stiffness in extension. It must be observed that the flexor tendons possess greater strength than the extensors, and this fact is responsible for recovery of flexion. During the first critical days, splinting the finger in extension prevents the protective attitude of flexion and in the author's experience has produced a better overall

result as regards correction of the flexion contracture. Following removal of the dressings at seven to ten days, active finger exercise is started. Close supervision, encouragement and reassurance are mandatory during this phase of recovery. In cases in which joint contracture is a problem, stretching with a dynamic splint during this phase of recovery may be helpful.

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CLUSTER MIGRAINE

"The crux of the cluster migraine is that it is predictably unpredictable, and the effectiveness of any prophylactic treatment that you use will depend in general upon the time you use it. If you use it in the fifth or sixth week, and the series or the cluster lasts for six weeks, you'll be a hero until the headaches start 18 or 24 months later. If you use it in the beginning, you'll have a stormier period. They've been known to stop as suddenly and for as unexplained reasons as they occur."

—ARNOLD P. FRIEDMAN, M.D., New York City
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